Specyficzne problemy pielęgnacyjne osób z zespołem Mayera-Rokitańskyego-Küstera-Hausera

(Specific problems of nursing patients suffering from Mayer-Rokitansky-Küster-Hauser Syndrome)

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Streszczenie - W pracy przedstawiono opis przypadku chorej z zespołem Mayera-Rokitansky'ego-Küstera-Hausera. Scharakteryzowano czynniki etiologiczne, obraz kliniczny, diagnostykę i metody leczenia. Szczególną uwagę zwrócono na specyficzne problemy pielęgnacyjne osób z tym zespołem.

Słowa kluczowe - zespół Mayera-Rokitańskyego-Küstera-Hausera, pielęgnacja chorych.

Abstract - Introduction. This paper presents a case description of a woman suffering from Mayer-Rokitansky-Küster-Hauser Syndrome. The etiological factors, the clinical presentation as well as the diagnostics and methods of treatment were scrutinized in this paper. Special attention was devoted to specific problems of nursing patients suffering from that syndrome.

Key words - Mayer-Rokitansky-Küster-Hauser Syndrome, nursing patients.

I. INTRODUCTION

The first descriptions of female sex organ malformations that could indicate a congenital absence or underdevelopment of vagina and uterus date back to ancient times. However, the cases described then are different from the modern-day clinical presentation of the syndrome. It is believed that Avicenna, who lived in the period of 980 to 1037, provided a foundation for the development of studies on this rare malformation of female genital system. Avicenna, or correctly: Abu Ali Ibn was a Persian doctor, philosopher, biologist and poet.

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He is considered one of the most versatile Arab scholars. His study was followed by research on vagina and uterus malformation published by different scientists.

In the 19th century August Franz Joseph Karl Mayer (1787 - 1865), Adolf Kussmaul (1822 - 1902) and Karl Rokitansky (1804 - 1878) explored the topic. They have presented cases of similar female condition [1].

Yet, it was not until 1910 that a German surgeon, Küster described the MRKH syndrome on the basis of studies conducted worldwide. The name itself, being derived from the last names of subsequent researches interested in the matter: August Mayer, Karl von Rokitansky, Herman Küster and Georges Hauser, is as follows: Mayer-Rokitansky-Küster-Hauser Syndrome (MRKH)[2].

II. ETIOPATHOGENESIS AND EPIDEMIOLOGY

MRKH syndrome is caused by embryonic disorders of internal genital organs. This malformation is a result of the arrest of development of absence of Müllerian ducts. That takes place around the fifth week of foetal life. The absence of Müllerian ducts makes vagina and uterus fail to be formed.

The frequency of the syndrome occurrence is estimated to be 1:4000 – 1:5000 out of living female neonates. It is worth mentioning that most women have standard karyotype. Sometimes MRKH co-occurs with other congenital defects. Those are related to urogenital system and include predominantly unilateral renal agenesis, ectopic kidney, often co-existing with heart and osseous system defects [3, 4].

III. THE CLINICAL PRESENTATION OF MAYER-ROKITANKY-KÜSTER-HAUSER SYNDROME

The most usual symptom of MRKH is the absence of menstruation. The affected women are not characterized by improper body structure or abnormal psychophysical development. Nipples and hair are correctly developed as related to the physical development stage on the Tanner scale. No sex-related disorders or androgynous features are observed. Genetic examinations indicate that the female karyotype is correct. No disorders are found in hormonal tests, either [5].

IV. IDENTIFICATION

The congenital absence of vagina and uterus is indentified on the basis of gynaecological examination and ultrasound test of the organs in small pelvis. Invasive diagnostic methods such as laparoscopy should be used in the cases of patients who suffer from recurrent abdominal pain [6].

During gynaecological and rectal examination no uterus is detected. Vulva is correctly developed, but vagina is absent or only a minor recess (1-2 cm) with a hymen [5].

V. CASE STUDY

A patient, W. G., 17 years of age, visited clinic K. in a primary care institution. She was alarmed by her lack of menstruation despite her advanced age.

The patient was born in 1992. W.G. was a first child of a 20-year-old woman and a 24-year-old man. Her mother did not smoke or drink alcohol when she was pregnant; she did not work, either. Her father, who smoked 30 cigarettes a day, worked as a driver. The general condition of both parents was good. W.G.'s birth was spontaneous and in due time. The newborn baby's weight was 3350g and her height was 50cm. Her Apgar score was 10.

W.G. underwent bilateral inguinal hernia as a neonate. As a child, she often had urinary tract infections.

During adolescence she developed all the tertiary sexual characteristics. At first she was not anxious about her lack of menstruation because she knew that women in her family started menstruating late.

Physical examinations

In the gynaecological examination, the external genital organs were found to be normal. No uterus was found during the rectal examination.

In the ultrasound examination of the abdomen, ovaries with scarce, small follicles without the dominant one were noted. Right ovary was 28 mm by 16 mm, whereas the left one was 32mm by 17mm. Fallopian tubes were present as well. There was no uterus.

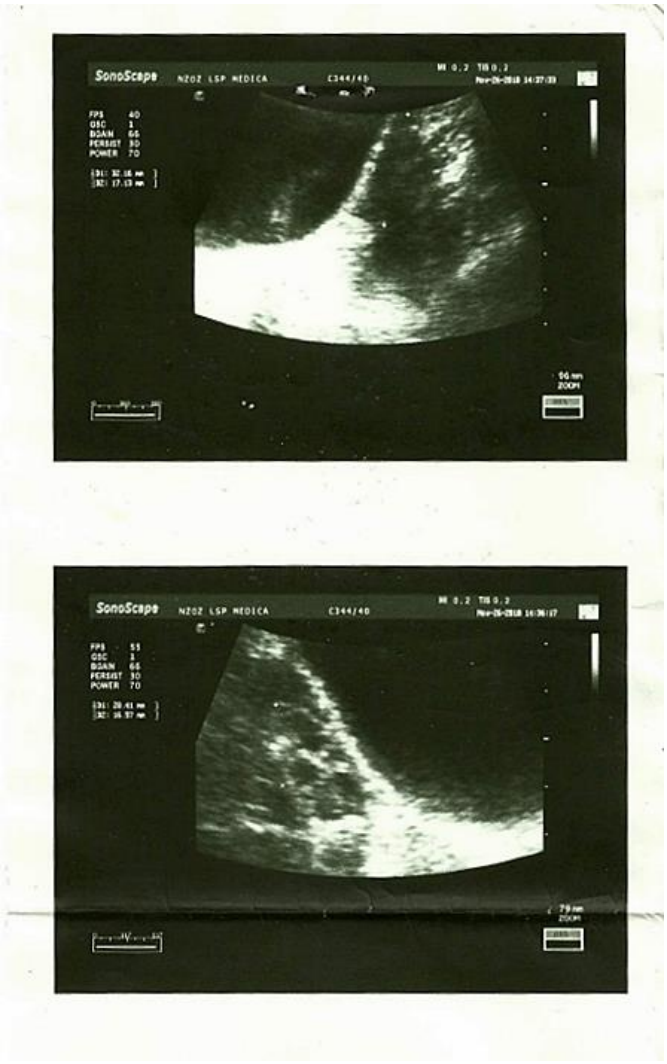

Fig. 1 The abdominal ultrasound examination result of W.G.

In one of the laboratory tests carried out the levels of the following hormones were measured: TSH, FT4, estradiol, LH and FSH. The levels of those, except for TSH, were normal. The TSH content in serum was 0,333 uIU/mL (normal: 0.4-4.0 ulU/mL).

The specificity of the problem

W.G. talks much of her problems and fears. Her major concern is whether or not she will be able to start a family with a partner and whether the potential partner could understand her

situation as a woman and a wife. W.G. does not accept the situation she found herself in, which makes her moody.

She is very much afraid of loneliness; in spite of that fact, she isolates herself from the people she knows. She confesses that sometimes she finds it difficult to understand herself. The hopelessness she feels makes her miserable, inert and fretful. She sees no way out of the situation.

She is anxious about her first sexual intercourse. The fear of the pain she might experience during vaginoplastic surgery prevents her from committing to further diagnosis and treatment. The impossibility of pregnancy and having biological children is a big problem for W.G. Although medical technology makes in-vitro method available if a surrogate mother is found, she does not accept this solution because of her religious beliefs. Also the absence of menstruation makes her frustrated, as healthy women treat it as unwelcome, negative state.

W.G. no longer feels she is a woman in the true meaning of the word. As she is exposed to opinions on role of a woman in the society, her approach deteriorates. After all, little girls are brought up to be mothers and raise children. Her moodiness causes many misunderstandings in her family. As a result, she has conflicts with her parents, which makes the girl feel misunderstood. She doesn't know how she could vent her anger.

VI . DISCUSSION

The most serious consequences for a woman with MRKH syndrome are the impossibility of having biological children and vaginal sex intercourse. Being aware of that, these women lose self-confidence related to their sex.

An essential element of care provided to those women is cooperation with w psychologist. Psychologists need to be involved in four critical periods, which are: the diagnosis of the malformation, vaginoplastic surgery (formation of vagina), entering a relationship with a man and starting a family.

It is important that upon the diagnosis of the syndrome the patient should be referred to additional examinations in which other defects could be discovered [3].

A woman should be informed about the malformation in an atmosphere of care and support. The way she learns about it has a great impact on the patient later on. It is crucial that during the same conversation the patient is told about her MRKH syndrome, doctors should emphasize that there is a possibility of becoming a biological mother using the in-vitro fertilization method and having a surrogate mother for the pregnancy period.

At present, several methods of surgical and non-surgical treatment of MRKH are available. Among them is the vaginoplastic surgery, which allows the patient to have her vagina surgically formed and makes sex life available. The treatment usually begins from an attempt of non-surgical vagina formation using the Franke's method [7]. That is available in the cases of patients whose vaginal recess exceeds 2,5cm– 3cm. The non-surgical method consists in pressing the recess with dilators, their sizes being gradually increased. The effectiveness of this method is as high as 85– 90% and is dependent on the patient's motivation as well as the support of the therapeutic team [3].

There are many methods of surgical vagina formation, with the three basic ones among them:

- A surgical method consisting in the formation of vaginal cavity between urethra with bladder and rectum,
- The Vecchietti procedure,
- vulvo-vaginoplasty.

The differences between each technique consist in the materials used in order to form the vagina, for instance: skin grafts, peritoneum, amnion, sigmoid, bladder and oral mucosa (cheek). It has to be underlined that virtually all patients who had vaginoplastic surgery need to have their vaginas widened using special dilators. Women should be informed about the necessity of that act earlier on. Using vaginal dilators followed by regular vaginal intercourse guarantee that an optimal size of vagina is formed and retained [6].

Among the numerous methods of vagina creation, each has its benefits and drawbacks. Different studies and modifications aiming at improving those methods are being undertaken. A vast majority of people who deal with patients suffering from the absence of uterus and vagina are of the opinion that a surgery should take place when the patient commences her sexual life; sometimes even later. The decision, however, is always for the patient to make [8].

The specific problems of the patient and nursing tasks needed to solve them

Listed here are the most significant problems encountered by the MRKH patient:

- Anxiety, doubts and insecurity about the obstacles to start a family,
- Fear of being rejected by her partner,
- Moodiness caused by the difficulties related to accepting the situation,
- Fear of loneliness together with isolation from people,
- Anxiety related to further diagnostics and treatment as well as first sexual intercourse,
- Impossibility of having biological children,
- Frequent conflicts with her parents and brother,

As most of the patients' problems are of mental nature, a nurse has to become a therapist, in a way. The nurse is here a basic psychotherapeutic unit. Such a role of a psychotherapist nurse is a new trend in the field. The nurse has a direct influence on the patient here. Very much depends on the nurse's personality, attitude and good manners. She is the one whose task is to introduce an air of trust and security.

In such a case it is the nurse's role to satisfy the patient's needs to be accepted, to belong somewhere, to be appreciated; also the needs of love and affection expressed by touching and friendly gestures. The person in charge of the patients should establish an individual therapeutic climate. The nurse's main task is convincing a girl that she is a perfectly normal woman. She must be reassured that she is no different than her peers and femininity is not measured in physical terms but in terms of spirit and personality.

A good mental state of the patient must be taken care of, the nurse should listen to her expressing her fears and clear her doubts about her condition. Nursing care also consists in putting the patient in touch with support groups, where patients are therapists to each other as they share one another's experiences [5]. The nurse should be able to cooperate with the patient's psychotherapist and her family. She should take interest in whether the patient's family has the knowledge on her illness and on the way she should be approached. If the answer is no, the nurse should skilfully teach them what is necessary to know and offer them advice.

What is more, a psychologist appointment should be recommended to the patient. Family therapy including the patient's partner is of utter importance. Such a therapy eases the tension and soothes conflicts. Family support and care are crucial for patients.

The psychological therapy should be aimed at minimizing the depression symptoms. The patient's participation in group therapy allows her not to feel lonely. That is beneficial, as most of MRKH patients tend to isolate themselves from friends and acquaintances in an attempt to have distance to life strategy or even to escape from the illness or the real world.

Knowing about these problems and about the patient's reluctance towards psychotherapy or support groups, the nurse should talk to the patient and encourage her to participate in a therapy. Nevertheless, it is important to remain tactful and considerate so as not to offend or discourage her. An approach of a good friend rather than a moralizer should be assumed. Empathy is helpful here. The final decision should always depend solely on the patient.

Special attention should also be paid to the necessity of the patient's partner being active during her psychotherapy. His role in supporting the young woman is unmatched. The relation between the patient and her partner should be based on dialogue, mutual understanding and affection. That could be a solution to another problem – the fear of vaginoplastic surgery and sexual intercourse. As far as having children is concerned, the nurse should talk to the patient about adoption and adoption centres.

Another task of the nurse is to point out the possibility of other congenital defects related to MRKH syndrome, due to which she should seriously consider further diagnostics.

All in all, in order to help MRKH patients not only immense knowledge on the part of the nurse is required. The cooperation of the therapist, the family and the patient's partner is also essential.

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